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Case Report

Targeted palliative endovascular embolization of a glomus jugulotympanicum tumor for refractory Jacobson's neuralgia: A case report

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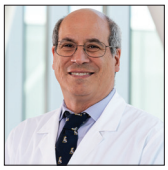

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ABSTRACT

Background: Glomus tumors around the jugular foramen and inner ear can have variable presentations, including lower cranial nerve palsies, tinnitus, hearing loss, or palpable neck mass. In general, these tumors are benign paragangliomas with the definitive treatment consisting of radiosurgery or surgery. Endovascular embolization can be added as a critical adjunctive therapy to reduce the tumor vascularity before surgical resection. We present the first case of a glomus jugulotympanicum presenting with a severe otalgia-dominant form of glossopharyngeal neuralgia, Jacobson's neuralgia, that was resistant to radiosurgery and relieved successfully by targeted endovascular embolization.

Case Description: A 51-year-old female presented with worsening right-sided lancinating ear pain radiating into the jaw and neck, exacerbated by brushing her teeth or any pressure on the skin – consistent with glossopharyngeal neuralgia, Jacobson's variant. Imaging revealed a dumbbell-shaped heterogeneously-enhancing mass in the middle ear cavity extending through the jugular foramen consistent with a glomus jugulotympanicum tumor. After treatment with single-fraction stereotactic radiosurgery, the neuralgia continued to worsen despite medical management and significantly impacted the patient's quality of life. After a multidisciplinary discussion, we performed targeted endovascular embolization of the tumor as palliative therapy. The patient subsequently reported complete relief of neuralgia and full resolution of tinnitus after the embolization procedure, remaining pain free at 20 months follow-up.

Conclusion: Targeted endovascular embolization may serve as a safe and potentially palliative option for refractory Jacobson's neuralgia induced by a glomus jugulotympanicum tumor.

Keywords: Embolization, Glomus, Glossopharyngeal, Jacobson, Neuralgia

INTRODUCTION

Glomus tumors are rare benign neoplasms originating in the head and neck that may be associated more commonly with hearing loss or tinnitus. Lower cranial nerve palsies are usually linked to glomus tumors as a result of tumor expansion or as a post-treatment complication.

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However, pain syndromes and neuralgia are not reported in relation to glomus tumors around the jugular foramen and inner ear.

Glossopharyngeal neuralgia is severe pain in the distribution of the ninth cranial nerve and represents 0.2–1.3% of the facial pain syndromes.[3,23] The most common form is the ear type – also known as Jacobson's neuralgia – with predominant otalgic symptoms.

In general, the treatment of glomus tumors primarily focuses on surgical resection or stereotactic radiotherapy. Before definite treatment, endovascular embolization is commonly used to reduce tumor vascularity and potential complications.

Here, we present the first case of glomus jugulotympanicum presenting with refractory Jacobson's neuralgia that was resistant to radiosurgery and relieved successfully by targeted endovascular embolization.

CASE PRESENTATION

History of present illness

A 51-year-old female presented with paroxysmal right-sided lancinating ear pain radiating into the jaw and neck, exacerbated by brushing her teeth or any pressure on the skin – consistent with glossopharyngeal neuralgia, Jacobson's variant. In addition, the patient had significant tinnitus over the same period. The patient had a normal exam with intact lower cranial nerves and no bradycardia. On otolaryngology evaluation, a mass behind the tympanic membrane was reported, along with mild sensorineural hearing loss in the right ear on the audiogram.

Diagnosis

Imaging revealed a dumbbell-shaped heterogeneously-enhancing mass located in the middle ear cavity and extending extracranially through the jugular foramen. The tympanic part of the mass measures 6 × 4 × 3 mm, while the jugular extension measures 15 × 15 × 15 mm with mild bony involvement and compression of the jugular bulb [Figure 1]. In addition, there was a slight increase in dopamine levels at the catecholamine screening.

The aforementioned clinico-radiological characteristics were most compatible with a jugulotympanicum paraganglioma. The tumor can be classified as type-II according to the Glasscock-Jackson classification (Glomus tympanicum Type II: Tumor filling the *middle ear*, and Glomus jugulare Type II: Tumor extending to the *internal auditory canal*, with or without intracranial extension).[13]

Stereotaxic radiosurgery (SRS)-treatment

Considering the tumor location, extension, and the patient's wishes not to undergo any open surgery, the patient underwent linear accelerator (LINAC)-based SRS 7 months after the symptoms' onset. The patient was treated with a single-fraction of 1500 cGY at the 78% isodose line with a conformity index of 1.55 and homogeneity index of 1.3 utilizing 4 non-coplanar arcs [Figure 2].

Post-SRS status

After treatment with stereotactic radiosurgery, the neuralgia remained refractory to any additional medical management, with the pain increasing to an 8 of 10 on the numerical rating scale, thus significantly impacting her daily life. The patient remained refractory to additional conservative management with a combination of gabapentin and topiramate, eventually failing a trial of Trileptal as well.

Endovascular-treatment

Given this refractory nature of the neuralgia, and based on a multidisciplinary discussion, it was decided that a targeted endovascular embolization of the superior portion of the tumor be performed as palliative therapy for her pain 15 months after the onset of symptoms.

The patient underwent targeted particle-based embolization under general anesthesia. Given the concerns for cranial nerve injury, monitoring was achieved of the lower cranial

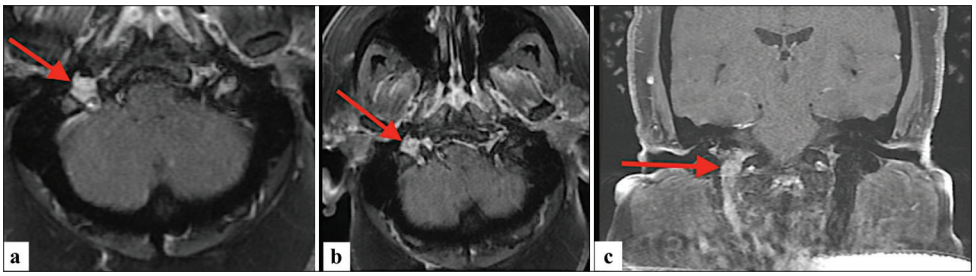

Figure 1: (a and b) Cranial magnetic resonant imaging T1 with contrast, axial view, and (c) coronal view demonstrating the tumor location (red arrow) within the right tympanic cavity, jugular foramen, and its extracranial extension.

nerves including VII, VIII, IX, X, and XII bilaterally. Tumor blush was noted from the right neuromeningeal trunk of the ascending pharyngeal artery through superselective microcatheter injection. A trial of amytal (25 mg) followed by lidocaine (20 mg) was slowly instilled intra-arterially with careful monitoring of all cranial nerves to assess for signal changes. No changes were detected. Therefore, 250–350 μm polyvinyl alcohol (Contour@; Boston Scientific Corp., Fremont, CA, USA) particles opacified with Omnipaque were injected into the targeted territory under biplane subtracted fluoroscopic guidance, avoiding any opacification of any territory beyond the tumor blush and any reflux. Interval acquisitions were performed, and the microcatheter was removed when the branches of the right neuromeningeal trunk were no longer opacified, consistent with proper embolization of the tumor. No tumor blush was noted on follow-up injections. Control monitoring remained unchanged, and the patient emerged from embolization with complete resolution of pain [Figure 3].

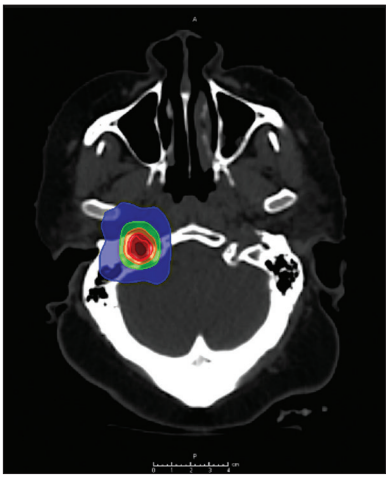

Figure 2: Stereotaxic radiosurgery using single fraction procedure targeting the present case of the right-sided glomus tumor.

Outcome

The patient reported complete resolution of neuralgia and tinnitus after the embolization procedure and continues to be pain-free at 20 months follow-up. Follow-up magnetic resonant imaging demonstrates some mild reduction in overall tumor volume.

Review of the literature

An extensive literature search was performed following the PRISMA guideline.[19] PubMed, Scopus, ScienceDirect, and Google Scholar were searched using the combination of the Boolean operators "OR" and "AND" and the search terms: "Glomus," "Paraganglioma," "Neuralgia," "Ear pain," and "Glossopharyngeal."

A total of 338 articles were retrieved from the literature. After the removal of 55 duplicates, the exclusion of articles was as follows: Review articles (n = 199), cadaveric/animal studies (n = 4), no glomus tumor (n = 54), and no glossopharyngeal neuralgia (n = 26). Consequently, there were no reported cases of glomus jugulotympanicum associated with Jacobson's neuralgia and treated with selective endovascular embolization.

DISCUSSION

Several types of glossopharyngeal neuralgia have been reported, including pharyngeal-type, cardiovascular-type, and ear-type.[5] Jacobson's neuralgia – also known as ear-type or tympanic – is the most common form of glossopharyngeal

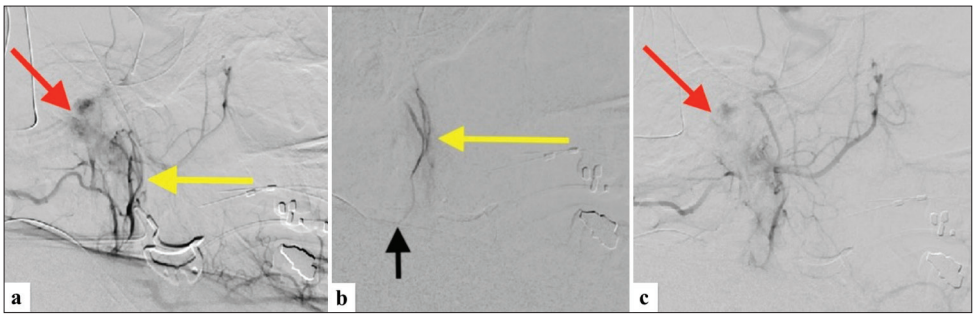

Figure 3: Endovascular treatment for the right-sided glomus tumor. (a) Pre-treatment selective microcatheter injection demonstrating the tumor blush at its superior pole (red arrow) being supplied by the jugular branch of the neuromeningeal trunk of the ascending pharyngeal artery (yellow arrow). (b) Targeted microcatheter, particle-based embolization of the more distal portion of the neuromeningeal trunk (yellow arrow) (black arrow). (c) Post-embolization injection demonstrated significant reduction of the tumor blush at its superior pole, presumptively the location where the greatest mechanical compression of Jacobson's nerve in the limited space at the jugular foramen (red arrow).

neuralgia that is manifested in our case.[7] In such cases, the pain markedly predominates in or remains confined, to the ear. Glossopharyngeal pain, especially coupled with pulsatile tinnitus, can be particularly debilitating and have a large impact on quality of life. In the present case, the patient had difficulty working, drinking, eating, and sleeping due to the severity of the pain. At one point, this patient had suicidal ideations secondary to her combination of symptoms.

Appreciating the anatomy of Jacobson's nerve helps us better understand the symptoms and triggers of our presenting case. Classically, Jacobson's nerve emerges from the inferior ganglion of the glossopharyngeal nerve and is considered the first branch of the ninth cranial nerve after having passed the jugular foramen [Figure 4].[6,9,21,24,25]

The literature described the etiology of glossopharyngeal neuralgia as idiopathic in the majority of clinical scenarios. Secondary glossopharyngeal neuralgia is extremely rare and can be attributed to infections or inflammations,[1,4] vascular lesions,[18,20] trauma,[11] elongated styloid process,[17] neoplasms such as meningioma, invasive nasopharyngeal carcinoma,[10] lymphadenoma,[15] tongue carcinoma,[2] osteoma foramen lacerum,[10] and neurofibroma[22] based on the current literature. Our case is the first to report the unusual presentation of glomus tumor as glossopharyngeal neuralgia.

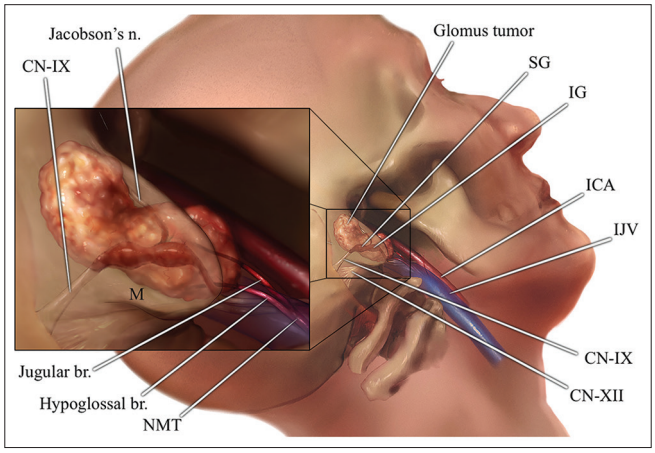

Figure 4: Illustration of the jugulotympanicum tumor and its presumptive relationship to Jacobson's nerve and its vascular supply from the neuromeningeal trunk of the ascending pharyngeal artery. The inset represents a magnified view of the relationship between two of the major distal branches of the neuromeningeal trunk (Hypoglossal and jugular branches) with respect to the tumor's location and to that of Jacobson's nerve. Given the confined space of the region at the level of the jugular foramen with the presence of the tumor, mechanical compression of the tympanic nerve, originating from the inferior ganglion, may contribute to Jacobson's neuralgia in this case. br. branch, CN-IX: Glossopharyngeal nerve, CN-XII: Hypoglossal nerve, ICA: Internal carotid artery, IG: Inferior ganglion, IJV: Internal jugular vein, M: Mastoid, n: Nerve, NMT: Neuromeningeal trunk, SG: Superior ganglion.

Glomus jugulare and tympanicum tumors are rare, highly vascular tumors with a reported incidence of 1 case per 1.3 million persons and represent 0.6% of all cranial neoplasms.[12] These tumors arise from various structures: Glomus jugulare tumors arising from paraganglion cells located in the adventitia of the jugular bulb; glomus tympanicum tumors arising from similar cells along Jacobson's nerve, the tympanic branch of the glossopharyngeal nerve, or Arnold's nerve, the auricular branch of the vagus nerve.[14] The paragangliomas in the head-and-neck region are often non-secretory; symptoms such as tremors, palpitations, headache, and excessive sweating are not commonly observed.[27] Up to 1–3% of glomus tumors can secrete catecholamines,[8] as was seen in this case.

Management of glomus jugulare and tympanicum tumors is challenging due to their rarity, increased vascularity, difficult anatomic locations, and advanced stages at presentation and diagnosis. When associated with devastating symptoms such as glossopharyngeal neuralgia, treatment should involve a multidisciplinary team of neurologists, neurosurgeons, and otorhinolaryngologists for early detection and improved outcomes. Conventionally, surgical resection has long been the standard of care for these tumors, but due to their high vascularity and the frequent involvement of the lower cranial nerves, surgery can carry significant risks. Radiotherapy has emerged as an alternative treatment for these tumors due to its relatively low-risk profile. However, the efficacy of radiosurgery in the immediate management of symptoms related to jugulotympanicum paraganglioma is limited as a result of the slow reduction in tumor size. This often results in patients experiencing ongoing tinnitus and cranial nerve deficits for months following radiosurgery, with some not experiencing full resolution and few experiencing a worsening of symptoms.[12]

The adequate stimulus for eliciting an attack of pain in such a patient seems to be a distortion of skin or mucosa. It has been supposed that afferent discharge arising from tactile receptors may play an important role in initiating pain paroxysms as a presumptive mechanism for neuralgic pain.[16] The long-term effectiveness and improvement in tinnitus, in our case, discourage the placebo effect of the management. One presumptive explanation of why the pain improved could be attributed to the removal of pulsatility against the nerve, hence reducing the severity of pain as is seen in the cases of microvascular decompression for tic douloureux. Another potential explanation is that the devascularization of Jacobson's nerve renders the region hypesthetic, for which the patient has no complaints during the follow-up.

In light of the challenges associated with surgical resection and the delayed treatment effect of radiosurgery, and given the debilitating and refractory nature of symptoms in this patient, endovascular embolization was adopted as the best treatment option for this case. This illustrates the principle of a changing "game-plan" in the context of a team-based

and patient-centered approach. Endovascular embolization involves the selective occlusion of blood vessels supplying the tumor, either resulting in the devascularization of the adjacent nerve supply or reduction of the pulsations or vascular mass of the tumor, hence reducing the mechanical impingement on Jacobson's nerve with subsequent symptom relief. When utilized as the sole treatment modality, embolization has been shown to have high recurrence rates for these tumors.[27] Large clinical trials comparing the efficacy of embolization to surgery or radiosurgery are not available and would be recommended as a future direction toward minimally invasive interventions and patient-centered care.

While embolization carries its risks, it presents a promising avenue for patients with complex Jugulotympanicum paraganglioma who have failed conservative management. The patient presented herein was experiencing progressively severe neuralgia that was refractory to standard medical treatment, which affected her activities of daily living. As a palliative effort to control pain, great care and planning were used in performing this targeted embolization. Regardless of the challenges, successful embolization of the tumor was achieved, and the patient exhibited complete relief of pain and resolution of tinnitus.

In this report, we described an unusual presentation of jugulotympanicum paraganglioma as Jacobson's neuralgia. In addition, the neuralgia was refractory to medical treatment and persisted after radiosurgery, with a significant impact on the patient's quality of life. A palliative targeted endovascular embolization was performed based on a multidisciplinary team decision and considering the patient's wishes and resulted in complete resolution of the patient's symptoms with maintenance of pain-free status on follow-up. The outcome of this report should be received with caution and further studies on this rare association would be recommended to improve our understanding of the best treatment of this disease entity. In addition, this report highlights the crucial role of the patient-centered and multidisciplinary team-based approach to improve the quality of care.

CONCLUSION

Targeted endovascular embolization can serve as a safe and potentially palliative option for refractory Jacobson's neuralgia induced by glomus jugulotympanicum tumor.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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